P.056

Optimizing IVIg Use for Neuromuscular Conditions in British Columbia, Canada – Targeting High and Chronic User Groups

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Background: Neuromuscular conditions account for 1/3 of IVIg use in BC and costs over \$10 million annually. Since 2013, the BC Neuromuscular Review Panel has developed diagnostic and treatment algorithms for the use of IVIg. A framework was created to review high dose and chronic users. Methods: Utilizing Central Transfusion Registry data, all patients treated with IVIg for approved neuromuscular conditions (CIDP, MG, MMN) since April 1. 2013 were identified. Annual cohorts for patients using higher than usual dose and chronic use (>3 years) were established, and evaluated annually. Patient specific recommendations were made. Results: The initial cohort identified 38 high users of 377 patients receiving IVIg. 27 appropriate, 9 "not appropriate". Subsequent cohorts showed a decrease in number of patients receiving inappropriate IVIg doses. In BC there has been a 36% increase in neuromuscular patients treated with IVIg (377 in 2013/14 to 512 in 2016/17). Despite this, IVIg the program has effectively reduced the annual grams/patient from 516 gm/patient in 2013/14 to 489 gm/ patient in 2016/17. Conclusions: The BC Neuromuscular IVIg Review confirms that the majority of IVIg use is appropriate. Following yearly cohorts of chronic and high dose users helps optimize IVIg use, which may lead to improved patient care.

P.057

Multidisciplinary Care for Optimal Management of Complex Nerve Injuries In Canada

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Background: Recent advances in management of peripheral nerve injuries is leading to a paradigm shift in the treatment of Canadian patients. Multi-disciplinary care models provide diagnostic, surgical and rehabilitative consultations within a single clinical encounter. Involvement of allied health care professionals has been shown to improve outcome. The purpose of this study was to ascertain the distribution and composition of multidisciplinary teams, and identify regional disparities. Methods: Representatives from clinics across Canada were invited to participate in a survey at the Annual Canadian Peripheral Nerve Symposium in London, Ontario in November 2019, with telephone follow up. Results: Delegates from 17 programs responded to the survey (12 academic centre and 5 community setting). Program provides electrodiagnostic testing, neuromuscular, rehabilitation and surgical assessment. Access to the following services was reported: occupational therapy=53% (9/17), physiotherapy 29% (5/17), research assistant=17% (3/17), social work=12% (2/17), mental health=6% (1/17). Conclusions: Complex nerve injury clinics are being established

throughout Canada. Allied health care and research support are limited in many multi-disciplinary complex nerve injury programs. There is variable access, likely resulting in disparities in patient care across Canada. This data will be valuable for lobbying for resources for resources to improve the care of these complex patients.

P.059

Results From the Randomized and Open-Label Periods of the CENTAUR Trial of Sodium Phenylbutyrate and Ursodoxicoltaurine in Amyotrophic Lateral Sclerosis (ALS)

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Background: An oral, fixed-dose sodium phenylbutyrate-ursodoxicoltaurine (PB-TURSO) coformulation was evaluated in a multicenter ALS trial (CENTAUR). Methods: Adults with definite ALS, ≤ 18 months from symptom onset, (N=137) were randomized 2:1 to PB-TURSO or placebo for 6 months. Completing participants were eligible to receive PB-TURSO in the openlabel extension (OLE) (≤30 months). The primary efficacy endpoint in both periods was rate of ALS Functional Rating Scale-Revised (ALSFRS-R) total score decline. All-cause survival was analyzed July 2020 (longest follow-up, 35 months). Safety was assessed in both periods. Results: Over 6-month randomized treatment, mean ALSFRS-R total score decline was slower with PB-TURSO vs placebo (difference, 0.42 points/month; P=0.03). Participants receiving PB-TURSO in the OLE (continued or crossover from placebo) maintained or initiated functional benefit beyond 6 months of therapy. Mean hazard of death was 44% lower (P=0.02) in the original PB-TURSO group. Overall adverse event (AE) incidence was similar, though early (week \leq 3) gastrointestinal AEs were more frequent during initial exposure to PB-TURSO (randomized period or OLE). Conclusions: PB-TURSO resulted in superior retention of function in the randomized period. Longterm OLE results support functional benefits of early vs delayed therapy and of sustained treatment. Survival was longer in the original PB-TURSO group after nearly 3 years.

P.060

Immunoglobulin Use For Neuromuscular Conditions: Updating British Columbia Provincial Guidelines Through Focused Literature Review

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Background: Immune-mediated neuromuscular conditions often cause significant disability and may require ongoing immunomodulating therapies such as immunoglobulin (Ig). Ig use in several neuromuscular conditions such as Chronic Inflammatory Demyelinating Polyneuropathy (CIDP) is supported by robust evidence, however Ig is increasingly used for other disorders.